Orthostatic Intolerance/OI

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DECEMBER 2017
Behavioral Objectives: Session 5

The learner will be able to:

- Measure **orthostatic intolerance** in a clinic visit using the 10 min Lean Test, or equivalent bedside testing
- Use the test results to formulate a supportive treatment plan.
ME/CFS Clinical Diagnostic Criteria:

**CORE/REQUIRED criteria:**  *Must be mod-severe and frequent (present >50% of the time)*

1) Impaired function related to exhaustion/fatigue
2) PEM: Post Exertional Malaise
3) Unrefreshing sleep
4) A. Cognitive impairment and/or
   B. Orthostatic intolerance

*Other common symptoms, not core:* Chronic widespread pain, Immune manifestations (allergy, inflammation, sensitivities), Infection manifestations, neuroendocrine manifestations
Orthostatic intolerance (OI) is the development of symptoms while standing upright that are relieved by reclining.
Orthostatic Intolerance

Orthostatic Intolerance/Autonomic Dysfunction/POTS (IOM Report, Chap 4, pp. 107-119)

1) Cerebral under-perfusion
   ◦ lightheadedness, fainting, impaired cognition, disorientation, headaches, visual changes, unusual neurologic symptoms, exhaustion

2) Peripheral cardiovascular signs
   ◦ Sympathetic nervous system activation---palpitations, nausea, abdominal and chest discomfort, facial pallor, cold hands and feet, anxiousness, shortness of breath, sweating, tremor...

Worsened by heat, dehydration, prolonged standing, deconditioning and weakness, and immediately after exercise
Symptoms of OI

**ACUTE OI**
- Fainting, lightheadedness
- Altered vision (blurred, double vision, tunnel vision)
- Anxiety
- Fatigue and weakness
- Headache
- Heart palpitations, heart pounding/racing
- Shortness of breath, hyperventilation
- Tremor

**CHRONIC OI**
- Nausea or low appetite
- Neurocognitive deficits, brain fog
- Pallor (pale)
- Sensitivity to heat
- Sleep problems
- Varied dizziness, vertigo
Orthostatic Intolerance/OI

- **Orthostatic hypotension:** a BP reduction of at least 20 mm Hg systolic or 10 mm Hg diastolic within the first 3 min of upright posture (American Autonomic Society and the American Academy of Neurology)

- **Postural Orthostatic Tachycardia Syndrome (PoTS):** the reproduction of orthostatic symptoms together with a **30 bpm** increase in HR, from supine to 10 min upright, or a HR of ≥120. Age 12 and 19 heart rate increase must be **40 bpm**

- **Neurally Mediated Hypotension (NMH):** synonymous with vasovagal syncope, neurocardiogenic syncope

- **Neurogenic Orthostatic Hypotension (NOH):** a distinctive and treatable sign of cardiovascular autonomic dysfunction. It is caused by failure of noradrenergic neurotransmission that is associated with a range of primary or secondary autonomic disorders, including pure autonomic failure, Parkinson’s disease with autonomic failure, multiple system atrophy as well as diabetic and nondiabetic autonomic neuropathies. (Metzler M 2012)
Orthostatic Intolerance/OI

Orthostatic Hypotension (OH)  ICD-10 I95.1

Conditions with no ICD-10 codes:
- Postural Orthostatic Tachycardia Syndrome (PoTS)
- Neurally Mediated Hypotension (NMH)
- Neurogenic Orthostatic Hypotension (NOH)

Dysautonomias in ICD-9 are now:
- Other disorders of the autonomic nervous system  G90.8,
- Disorder of the autonomic nervous system  G90.9
Assess and treat OI

Orthostatic intolerance is:

- Measurable (heart rate, blood pressure)
- Treatable or at least manageable.

OI can exist in someone who has low, normal or high blood pressure in the seated position.
Measuring orthostatic intolerance

Tilt Table test (not readily available or standardized is the “gold standard”)

Bedside orthostatic vital signs---Stand Test (1, 3, 5, 10 minute measures of BP and HR.

10 min “NASA” Lean Test

FitBit or other HR tracking devices can track heart rate as an indicator of exercise effort, but also an indirect measure of orthostatic intolerance
10 min informal stand test

19 year old male. BMI 18. Intake BP 110/64 and P 80

Became ill in 9th grade while training for cross country. Felt run down. Sick more often. Then IBS → Nausea and dizziness → Tension and migraines → Exercise intolerance. → Abdominal and chest pain → Couldn’t finish the year. Struggled with ups and downs sophomore, junior and senior year. Set off defiantly for college on his own...but returned

Heart Rate (HR) seated and relatively relaxed: **89 bpm**

- standing at 1 min 104 "it feels like I'm heavy; I feel light headed, weak"
- standing at 2 min 120
- standing at 3 min 113 "head hurting more, harder to concentrate"
- standing at 4 min 123 "now my leg muscles are hurting"
- standing at 5 min 115
- standing at 6 min 118 "hands and feet are definitely very heavy right now"
- standing at 7 min 117
- standing at 8 min 115
- standing at 9 min 120 “everything above is getting worse, blurred vision”,
- standing at 10 min **129** “starting to shake”

**HR increases 40+** Brain “checked out” Return for full NASA 10 min lean test.
10 min NASA lean test “prepped”

19 year old male. No medications in the last 24 hours and not wearing any compression clothing. He has stopped pushing water intake and so drinking a little less water than normal.

**Lying on bed at rest:**
- Supine 1 minute BP: 131/65  Pulse: 86  Pulse ox 98%
- Supine 2 minute BP: 131/65  **Pulse: 82**
- Supine 3 minute BP: 130/61  Pulse: 89  **PULSE PRESSURE: 69**

**Standing straight with shoulder blades against the wall and feet 6" from the wall**
- Standing 0 minute BP: 126/54  Pulse: 114  Feels “blood going down,” light headed, weak
- Standing 1 minute BP: 116/71  Pulse: 112  Pulse ox 95%
- Standing 2 minute BP: 121/82  Pulse: 100
- Standing 3 minute BP: 112/86  Pulse: 105
- Standing 4 minute BP: 118/85  Pulse: 107  Pulse ox 94% "Just worse" Starting to shake
- Standing 5 minute BP: 116/80  Pulse: 111
- Standing 6 minute BP: 115/85  Pulse: 121
- Standing 7 minute BP: 111/89  Pulse: 117 "Lack of concentration, getting headache, achy"
  Dependent rubor
- Standing 8 minute BP: 113/76  Pulse: 114
- Standing 9 minute BP: 112/79  Pulse: 123 "Feels like I'm breathing heavily"
- Standing 10 minute BP: 114/86  Pulse: 128  **PULSE PRESSURE: 28**

SBP dropped from 130 to 111 (-19)  DBP 61---〉 54---〉 89  HR increased from 82 to 128 (+40)
10 minute NASA Lean test
10 min NASA lean test

58 year old woman with CFS. BMI 17. Unaware of her OI

Lying down on bed at rest:
Supine 1 minute BP: 114/76 Pulse: 75 Pulse ox 98%
Supine 2 minute BP: 112/78 Pulse: 75

Standing leaning shoulder blades against the wall:
Standing 0 minute BP: 111/86 Pulse: 89
Standing 1 minute BP: 118/80 Pulse: 90 Pulse oximeter 95% "Lightheaded"
Standing 2 minute BP: 120/92 Pulse: 92
Standing 3 minute BP: 120/98 Pulse: 93 "Tired"
Standing 4 minute BP: 121/98 Pulse: 94 "Trying to catch breath"
Standing 5 minute BP: 123/100 Pulse: 95 "Heavier breathing and the desire to sit"
Standing 6 minute BP: 124/90 Pulse: 97 Pulse ox 94%
Standing 7 minute BP: 116/52 Pulse: 98. "Feels very different but cannot explain it"
Standing 8 minute BP: 108/50 Pulse: 99 Pulse ox 92%
Standing 9 minute BP: 108/60 Pulse: 100 "Feeling hot, thirsty, blurry vision"
Dependent rubor (purple) of feet noted
Standing 10 minute BP: 95/50 Pulse: 100 "Need to lie down"

SBP -19. DBP -28. HR +25 bpm. Symptoms and exam findings match changes in VS.
32 year old woman with severe migraines, fibromyalgia, depression, dizziness. She has not taken any of her morning meds and is not wearing compression clothing today.

**Orthostatic Vital Signs/The NASA LEAN Test**

<table>
<thead>
<tr>
<th>Position</th>
<th>BP</th>
<th>Pulse</th>
<th>Pulse Pressure</th>
</tr>
</thead>
<tbody>
<tr>
<td>Supine #1</td>
<td>118/64</td>
<td>89</td>
<td></td>
</tr>
<tr>
<td>Supine #2</td>
<td>116/60</td>
<td>85</td>
<td>54</td>
</tr>
<tr>
<td>Standing</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Standing 0 minute</td>
<td>104/80</td>
<td>85</td>
<td></td>
</tr>
<tr>
<td>Standing 1 minute</td>
<td>108/74</td>
<td>119</td>
<td></td>
</tr>
<tr>
<td>Standing 2 minute</td>
<td>96/70</td>
<td>116</td>
<td></td>
</tr>
<tr>
<td>Standing 3 minute</td>
<td>108/75</td>
<td>Pulse: 123</td>
<td>Arms &quot;almost feel like they are tingling&quot;</td>
</tr>
<tr>
<td>Standing 4 minute</td>
<td>98/78</td>
<td>120</td>
<td></td>
</tr>
<tr>
<td>Standing 5 minute</td>
<td>96/73</td>
<td>123</td>
<td>Lightheaded and dizzy (as if she is spinning)</td>
</tr>
<tr>
<td>Standing 6 minute</td>
<td>91/73</td>
<td>125</td>
<td></td>
</tr>
<tr>
<td>Standing 7 minute</td>
<td>94/74</td>
<td>122</td>
<td></td>
</tr>
<tr>
<td>Standing 8 minute</td>
<td>96/74</td>
<td>122</td>
<td></td>
</tr>
<tr>
<td>Standing 9 minute</td>
<td>92/79</td>
<td>126</td>
<td>Increased lightheadedness, nausea</td>
</tr>
<tr>
<td>Standing 10 minute</td>
<td>93/80</td>
<td>120</td>
<td>Increased &quot;electrical buzz&quot;</td>
</tr>
</tbody>
</table>

**Summary:**

- **-27 mmHg drop in SBP** meets criteria for orthostatic hypotension (> 20 mmHg decrease)
- **+41 bpm increase in Heart Rate** meets criteria for POTS (>30 bpm increase)
37 year old professional woman with ME/CFS.
2-4 HUA/d. Sitting: BP 112/75. P-77

Lying down resting:
Supine: BP 99/68 P-68 pulse pressure: 31

Standing with shoulders against wall, feet 6” from the wall.
Standing at 0 minutes: BP 99/72 P-90
Standing at 1 minute: BP 90/74 P-100 mild weakness, heavy feeling in legs
Standing at 2 minutes: BP 101/74 P-94 dependent rubor, facial pallor
Standing at 3 minutes: BP 104/84 P-111 hands tingling pulse pressure: 20
Standing at 4 minutes: BP 104/83 P-101 nausea
Standing at 5 minutes: unable to measure
Standing at 6 minutes: BP 88/62 P-132 palpitations
Standing at 7 minutes: BP 94/64 P-115 palpitations, increased nausea
Standing at 8 minutes: did not register on B/P cuff
Tingling in face increased, tingling all over, sees “spots”, muted sounds, legs gave way, vision blacking out. We assisted her then to slide supine onto floor.

HR 68 → 132 (+64 bpm) just before near-syncope (near-fainting).
Baseline low BP, plus POTS and NMH
Central Nervous System (CNS)
- Brain and spinal cord
- Integrative and control centers

Peripheral Nervous System (PNS)
- Cranial nerves and spinal nerves
- Communication lines between the CNS and the rest of the body

Sensory (afferent) division
- Somatic and visceral sensory nerve fibers
- Conducts impulses from receptors to the CNS

Motor (efferent) division
- Motor nerve fibers
- Conducts impulses from the CNS to effectors (muscles and glands)

Sympathetic division
- Mobilizes body systems during activity ("fight or flight")

Parasympathetic division
- Conserves energy
- Promotes "housekeeping" functions during rest

Autonomic nervous system (ANS)
- Visceral motor (involuntary)
- Conducts impulses from the CNS to cardiac muscles, smooth muscles, and glands

Somatic nervous system
- Somatic motor (voluntary)
- Conducts impulses from the CNS to skeletal muscles
OI may be caused by:

- Internal bleeding
- Dehydration
- Becoming overheated (e.g. soaking in a hot tub)
- Emotionally stressful events---vasovagal syncope
- Prolonged bedrest
- Space travel and returning to earth’s gravity
OI may be caused by:

**DRUG SIDE EFFECTS:** diuretics, tricyclic antidepressants, blood pressure drugs, drugs for prostate disease (doxazosin, tamsulosin), Yaz birth control (drospirenone/ethinyl estradiol), nitroglycerin, alcohol...

**HEART:** Heart arrhythmias, heart valve failure, heart attack (myocardial infarction), cardiomyopathies (disease of the heart muscle)

**LUNG:** Pulmonary embolus, primary pulmonary hypertension

**CENTRAL NERVOUS SYSTEM:** Brain lesions, Parkinson’s, Multiple Sclerosis, Parkinsons...

**PERIPHERAL NERVOUS SYSTEM:** diabetic neuropathy, spinal cord injury, small fiber neuropathy (may underlie FM and/or ME/CFS)
Chronic OI may be related to:

Dysfunction of the autonomic nervous system
- Toxic causes (e.g. vincristine chemotherapy)
- Autoimmune disease (e.g. Sjogrens)
- Post-viral, post-infection syndromes
- Small fiber neuropathies

Orthostatic Intolerance and dysautonomia are commonly found in FM, CFS, IBS, and Ehlers-Danlos Syndrome (hypermobility syndromes)
Interventions for OI:

Recognize and **avoid** the common triggers

- Heat, getting overheated
- Dehydration
- Prolonged standing in place
- Prolonged sitting
- Prolonged bedrest *(confuses the ANS)*
- Muscle atrophy and weakness
  - Abdomen/core, legs
- Medications that cause/worsen OI
Interventions for OI:

External **compression** or internal **constriction** of blood vessels

- Compression socks, pants, sleeves, abdominal binder
  - COST: one time cost of purchase then free use
- **Midodrine**, a peripheral alpha-1 receptor agonist (stimulates the receptor).
  - Alpha-1 adrenergic receptors are in blood vessels of the skin, GI, GU and brain (but midodrine doesn’t cross the blood brain barrier)
  - Constricts arterioles and veins that have Alpha-1 receptors and raises blood pressure.
  - Short half-life requires 3x/day dosing, every 3-4 hours, for continuous benefit.
  - Goosebumps, scalp tingling, cold hands and feet are common
  - COST: 10 mg, #90, generic co-pay, $400/mo cash price
Interventions for OI:

Increase **volume** in the vasculature (blood vessels)

- Consume extra **water/fluids** to expand blood volume
  - >1 gallon or >2 liters daily as a foundation

- Increased **salt intake** helps retain water in the circulation and tissues
  - Must match the fluid intake. 2-5 gms daily (1/2 to 1 tsp)

- **fludrocortisone** 0.1 mg once or twice daily (sodium retention at the kidney)

- **desmopressin.** Only under *skilled* medical supervision. Reduces water loss by kidney. Recycles water rather than losing water in urine. Risk of hyponatremia. Also a vasoconstrictor.

- **Rapid water ingestion** (16 oz) helps reduce OI within 20 minutes (chugging)
Interventions for OI:

Control/cap the rapid heart rate response if helpful.

- Low dose beta blockers:
  - propranolol
  - metoprolol
  - atenolol

- Beta blockers can slow the HR too much, drop BP further, or block B2 receptors in the lung needed for asthma rescue with bronchodilators, which are B2 agonists (stimulate B2 receptors)
Interventions for OI:

Improve the muscular “pumps” for better venous return.

Muscular pumps = leg and abdominal muscles
Interventions for OI:

Exercise with a goal to increase:
- muscular strength in legs and trunk
- General circulation from light aerobic activity

Exercise tips
- **Drink 500 cc cold water** 20 minutes prior to exercise
- Wear **compression** socks, pants, abdominal binder during exercise
- Lie down, elevate feet and hydrate **immediately after** exercising
- **Lie down or sit** during exercise or movement activities
- Work gradually into the cardio (walking) by doing very short bouts
  - 2, 5, 10, 15 min
- **Exercise in water!**
  - Easier on joints and low back
  - Cool water helps vasoconstriction
  - warm water helps widespread pain
  - Hydrostatic pressure as water depth increases acts as “compression”
  - Swimming horizontal eliminates orthostatic stress

### Table: Water Pressure

<table>
<thead>
<tr>
<th>Depth of Water Column Vertical Feet</th>
<th>Water Pressure lbs. per sq. in.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>0.45</td>
</tr>
<tr>
<td>2</td>
<td>0.91</td>
</tr>
<tr>
<td>3</td>
<td>1.36</td>
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<tr>
<td>4</td>
<td>1.82</td>
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<tr>
<td>5</td>
<td>2.27</td>
</tr>
<tr>
<td><strong>6</strong></td>
<td><strong>2.73</strong></td>
</tr>
<tr>
<td>7</td>
<td>3.18</td>
</tr>
<tr>
<td>8</td>
<td>3.64</td>
</tr>
<tr>
<td>9</td>
<td>4.09</td>
</tr>
<tr>
<td>10</td>
<td>4.55</td>
</tr>
</tbody>
</table>
OI-friendly activities

Lying down or seated “movement” ideas:

- Restorative yoga
- Recumbent bike
- Weights or bands
- Planks

Water exercise
Clinical significance (OI):

Orthostatic Intolerance syndromes are diagnosable and treatable.

Substantial evidence base regarding these syndromes.

Improving OI improves symptoms and function and gives the patient more control over activity tolerance.
Orthostatic intolerance resources

https://www.youtube.com/user/OFFERUtah/videos

Melissa Cortez MD
◦ "Remaining Upright: Approach to Orthostatic Intolerance."

Craig Coleby MD
◦ “Redefining POTS so that Everybody Understands”

http://dysautonomiainternational.org/

http://www.dinet.org/

Wikipedia: Orthostatic Intolerance or dysautonomia

Medscape: Orthostatic Intolerance, Julian M. Stewart MD, PhD 2/2/15
◦ Drugs & Diseases > Neurology